Hereditary Hemorrhagic Telangiectasia with Pulmonary Arteriovenous Malformations and Embolic Strokes Treated Successfully with Video-assisted Thoracoscopic Resection

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Abstract

A 67-year-old hypoxic woman was admitted following two episodes of cerebral infarction. Based on the clinical presentation and radiological findings, a diagnosis of hereditary hemorrhagic telangiectasia was made and the cerebral ischemic complications were considered to have been caused by paradoxical embolizations related to pulmonary arteriovenous malformations (PAVMs). We performed video-assisted thoracoscopic surgery (VATS) and identified turbulent thrombi in one of the PAVMs that were capable of circulating systemically and inducing embolic strokes. The condition of the patient improved immediately following VATS. This case suggests that VATS may be a preferred therapeutic option in the treatment of patients with PAVM accompanying recurrent episodes of life-threatening complications such as strokes.

Key words: pulmonary arteriovenous malformation, hereditary hemorrhagic telangiectasia, cerebral infarction, video-assisted thoracoscopic resection

Introduction

Treating pulmonary arteriovenous malformation (PAVM) is essential, not only to improve hypoxemia, but also to prevent latent complications such as strokes and infections. Strokes are a life-threatening complication of hereditary hemorrhagic telangiectasia (HHT) that are presumed to occur due to paradoxical embolization related to PAVM. Transcatheter embolization, a standard procedure for treating PAVM, is recognized to be efficacious with a good safety profile (1, 2). Although transcatheter embolization is less invasive, the risk of stroke recurrence caused by the migration of the embolism still remains. We herein report the case of a hypoxic patient with recurrent episodes of strokes caused by PAVMs associated with HHT who was successfully treated with video-assisted thoracoscopic surgery (VATS).

Case Report

A 67-year-old Japanese woman was admitted to our hospital with muscle weakness in the right upper extremity and neck pain. The patient had been diagnosed with PAVM 22 years previously on chest radiography during a routine medical checkup. The patient’s clinical history at that time was remarkable for dyspnea on light exertion; however, her respiratory system and circulation were not evaluated further. She had experienced an episode of chronic recurrent epistaxis, and her close relatives also exhibited clinical symptoms associated with telangiectasias (Fig. 1).

On physical examination, the patient was afebrile and had a regular pulse of 80 beats/min and a blood pressure of 165/98 mmHg. There were telangiectasias present on her tongue, right first finger and anterior chest wall (Fig. 2A, B). In addition, auscultatory bruits were heard in the right posterior
feeding arteries larger than 3 mm in diameter and drainage veins located in the subpleural region (Fig. 3C, D, E, F).

In the laboratory analysis, the patient’s complete blood count, blood chemistry results and coagulation values were almost normal, except for a low-density cholesterol level of 154 mg/dL. An arterial blood gas analysis revealed a pH of

lower chest. On neurological examination, although the patient was fully alert and oriented, slight dysdiadochokinesis was observed in her left hand. Her National Institutes of Health Stroke Scale (NIHSS) score was zero (the highest possible score is 42).

Diffusion-weighted magnetic resonance imaging (MRI) revealed a right cerebellar infarction, and magnetic resonance angiography (MRA) revealed basilar arterial occlusion (Fig. 3A, B). Chest radiography revealed a nodule in the right upper lobe and abnormal shadows in the right lung base. Three-dimensional computed tomography (3D-CT) angiography of the chest revealed a 20-mm nodule in the right S2 segment, a 15-mm nodule in the right S5 segment and a 30-mm nodule in the right S9 segment, all of which had feeding arteries larger than 3 mm in diameter and drainage

Figure 1. The circles represent women and the squares represent men. The slashes indicate people who died. The gray squares and circles represent a history of epistaxis and arteriovenous malformation, respectively. The arrow indicates this patient.

Figure 2. Telangiectasias are present on the patient’s right first finger (A) and tongue (B). Small dot-like lesions are also observed (arrow).

Figure 3. A: An axial MRI scan obtained using diffusion-weighted imaging reveals a high-intensity signal in the right cerebellar infarction (white arrow). B: Magnetic resonance angiography (MRA) reveals occlusion of the basilar artery (white arrow). C: Chest radiography reveals a nodule in the right upper lobe (arrow) and abnormal shadows in the right lung base (arrowhead). D: Image acquired using three-dimensional computed tomography angiography reveals three pulmonary arteriovenous malformations located in the subpleural area of the right S2 (white arrow), S5 (white arrowhead) and S9 (black arrow) segments. E: Computed tomography shows a pulmonary arteriovenous malformation located in the subpleural area of the right S2 segment (white arrow). F: Computed tomography shows pulmonary arteriovenous malformations located in the subpleural area of the right S5 (white arrowhead) and S9 (black arrow) segments.
7.47, a arterial O₂ pressure (PaO₂) of 59 Torr and a arterial oxygen partial pressure (PaCO₂) of 34 Torr while the patient breathed ambient air. The shunt fraction was 19.6%.

On physiological examination, the result of a pulmonary function test was normal with a normal vital capacity (2.37 L, 98%) and forced expiratory volume in one second (2.02 L, 85%); however, a reduced diffusing capacity for carbon monoxide of the predicted value of 79% was observed. On electrocardiogram (ECG) and Holter ECG screenings, a sinus rhythm with no signs of arterial fibrillation was identified. Carotid duplex ultrasound revealed no indications of stenosis of the carotid artery or dissection of the vertebral artery. ECG revealed no thrombi in the left atrium, and the predicted pulmonary arterial pressure was 20 mmHg, thereby excluding a diagnosis of pulmonary hypertension. An endoscopic evaluation revealed several areas of gastric telangiectasias. A clinical diagnosis of HHT was made based on the consensus clinical diagnostic criteria known as the Curaçao Criteria (3): 1) spontaneous recurrent nosebleeds, 2) mucocutaneous telangiectasia (multiple at characteristic sites: fingertip pulps, oral mucosa or tongue), 3) visceral involvement (gastrointestinal, pulmonary, hepatic, cerebral or spinal arteriovenous malformation) and 4) a family history (first-degree relative affected).

Six days after the initiation of antiplatelet (aspirin, clopidogrel) and anticoagulation (Argatroban) therapy for cerebellar infarction, the patient presented with focal neurological symptoms such as left hemiparesis, left-sided sensory paralysis and dysarthria. Diffusion-weighted MRI revealed a new high-intensity signal in the region supplied by the right middle cerebral artery. MRA revealed occlusion of the right internal carotid artery and right middle cerebral artery (Fig. 4A, B). The NIHSS score was 16/42. Endovascular revascularization with thrombectomy for occlusion of the right internal carotid artery was performed. This intervention was effective, and the NIHSS score improved to 1/42 following thrombectomy.

In order to prevent any recurrence of cerebral infarction and improve the hypoxia, the patient underwent VATS. Three PAVMs located in the subpleural area of the right S2, S5 and S9 segments were excised successfully. During VATS, we were able to clearly observe all PAVMs and the turbulent thrombi in a PAVM located in the right S2 segment (Fig. 5). Pathologically, organizing mural thrombi were present in the resected PAVMs (Fig. 6). Marked relief of dyspnea was obtained immediately following VATS. An arterial gas analysis revealed a PaO₂ of 105 Torr, and the shunt fraction improved to 9.6%. There were no complications, and the two-year follow-up period was uneventful.

**Discussion**

In the absence of a normal capillary bed to filter particulate matter that traverses the lungs, the most frequently reported complications of PAVM are related to neurological sequelae such as strokes and brain abscesses. The use of transcatheter embolization is preferred in almost all patients with PAVM because it has a high success rate, is less inva-
Figure 6. The pathological findings of the pulmonary arteriovenous malformations show intimal thickening and organizing mural thrombi (arrows) that were observed during video-assisted thoracoscopic surgery.

In this patient, we performed VATS to treat the PAVMs relatively early following recurrent embolic strokes, and the procedure was successful without any surgery-related complications or recurrence of cerebral infarctions. There are several reasons why we chose VATS instead of transcatheter embolization for this patient. First, the number and location of the PAVMs were suitable for surgery; local resection during VATS was highly successful for three isolated and subpleural PAVMs. Temes et al. (4) reported that small PAVMs located in the subpleural region can be locally resected using VATS. Another reason why we chose VATS for the treatment of PAVMs was that the cerebral infarctions seemed to occur via paradoxical embolization related to the PAVMs. In fact, we observed some turbulent thrombi in one of the PAVMs that were capable of circulating systemically and inducing cerebral infarctions.

Transcatheter embolization with detachable balloons or coils results in the angiographic occlusion of the afferent vessels of the PAVMs. However, transcatheter embolization alone is not sufficient for the treatment of PAVMs with ischemic complications. Because there is a possibility that PAVMs contain thrombi, physicians must manage not only the afferent, but also the efferent vessels of the PAVMs in order to eliminate the risk of ischemic episodes. The acute periprocedural complications of transcatheter embolization include ischemic strokes and paradoxical embolization (2-4%) (5, 6). The appropriate method of treatment depends on the number and location of the PAVMs. Subpleural isolated PAVMs can be removed via local resection. In addition, smaller PAVMs and single efferent and afferent vessels are also suitable for surgery. In our case, all of the PAVMs satisfied these conditions. Therefore, we selected VATS (less invasive than thoracotomy) to treat the PAVMs in our patient. Both the efferent and afferent vessels of the PAVMs were ligated and successfully removed.

In conclusion, because transcatheter embolization is considered to be a very safe and effective intervention for PAVMs, the surgical indications can therefore be expanded to include patients with recurrent ischemic complications that are considered to be caused by paradoxical embolization related to PAVMs.

The authors state that they have no Conflict of Interest (COI).

References